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Pediatric Pial AVF

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A pial arteriovenous fistula (AVF) is defined by a simple direct communication between an artery and vein (figure 1). The first described case of diagnosis and treatment was by Walter Dandy in 1928, when he ligated a single feeder, which collapsed the enlarged venous drainage in an adult patient presenting with seizures [1]. Formal categorization and recognition of the pial Arteriovenous fistula as a distinct entity was described by Pierre Lasjaunias [2].

Pial arteriovenous fistulas are unusual lesions. In a review by Halbach, et al in 1989, 1.6% of their 320 AVMs were single hole fistula. [3] However, it would appear that they are more common in the pediatric group. In 1992, the combined series of cerebral AVF from the University of Western Ontario and UCLA and a review of the literature were reported. At that time, there were 13 fistulae identified in their combined group, and 35 identified from the literature. Overall, when the age could be determined, there were 27 in the pediatric group versus 10 in adults.

The clinical symptoms varied and 10% were found to by Asymptomatic. Of the 5 neonates, all presented with congestive heart failure. In infants, 6/11 had increasing head circumference, 3/11 had seizures, 4/11 had some degree of CHF, and 1/11 had haemorrhage. In children and adolescents, 7/11 had headache (with or without nausea and vomiting), 4/11 had ataxia or hemiparesis, 5/11 had seizures, and 1/11 had haemorrhage. In Adults, 4/10 had focal deficits, 3/10 had headaches, 1/10 had trigeminal neuralgia, 2/10 had seizures, and one had subarachnoid haemorrhage [4].

The most recent series and review of the literature was by Hoh in 2001. At their institution, adult patients presenting with haemorrhage predominated. But in the overall review, there were approximately more pediatric patients identified. Although the data are hard to extract, it seems that in adults, haemorrhage, headache and focal neurologic symptoms were all common presenting signs. In neonates, although haemorrhage can be seen, most present with CHF. In infants (1 month-2 yrs) symptoms due to hydrovenous disorders including: increasing head circumference, focal neurologic symptoms and increased ICP, haemorrhage and developmental delay all occurred in roughly equivalent frequency (figure 2). In children and adolescents, the hydrovenous disorder presented as focal signs, seizures and haemorrhage. Although these series may not have specifically looked for genetic disease, there were 8 patients with Hereditary Hemorrhagic Telangectasia (HHT), and one with Klippel-Trenaunay-Weber [5].

There is a predilection for Pial Arteriovenous fistula in HHT, and arteriovenous malformations (AVM) and multiple fistula are more

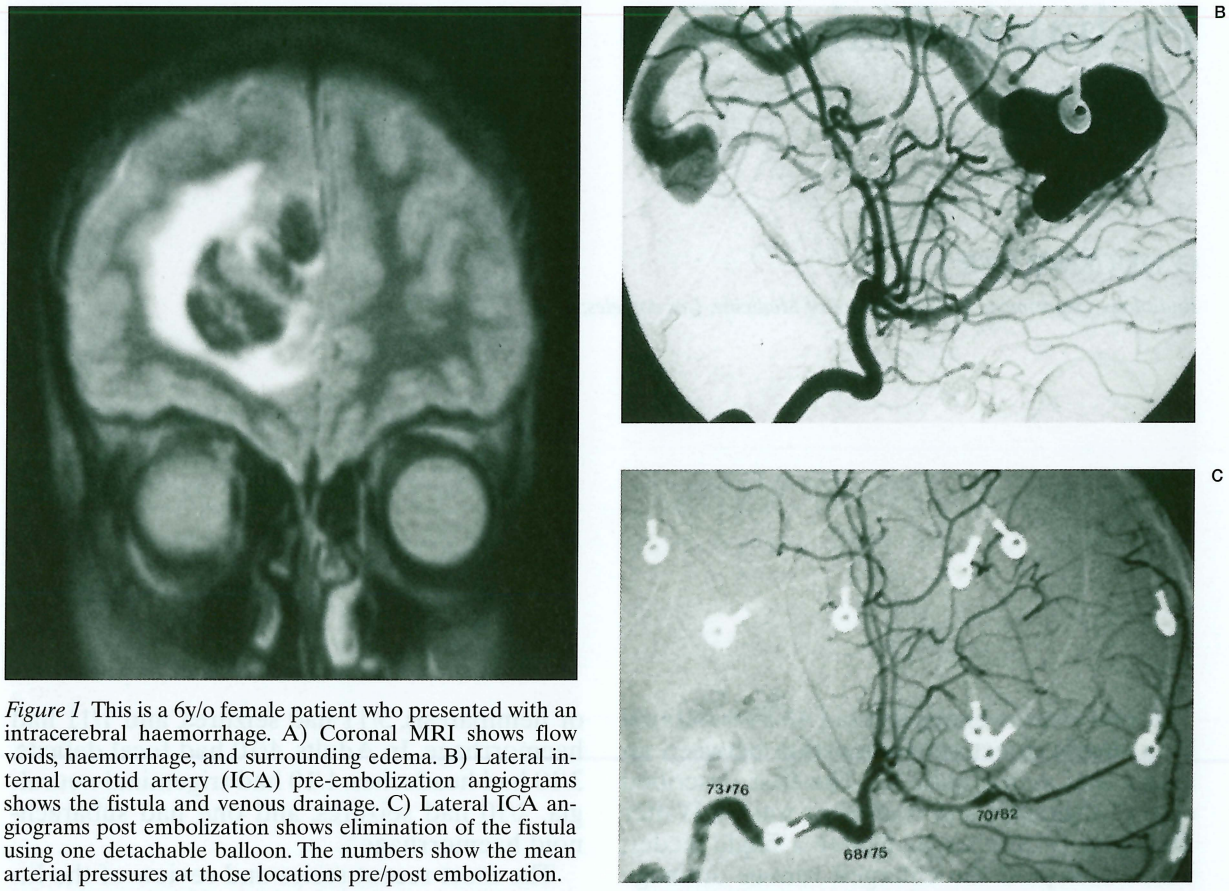

Figure 1 This is a 6y/o female patient who presented with an intracerebral haemorrhage. A) Coronal MRI shows flow voids, haemorrhage, and surrounding edema. B) Lateral internal carotid artery (ICA) pre-embolization angiograms shows the fistula and venous drainage. C) Lateral ICA angiograms post embolization shows elimination of the fistula using one detachable balloon. The numbers show the mean arterial pressures at those locations pre/post embolization.

common in the HHT population. In HHT, 8% of patients will have central nervous system (CNS) AVMs, and 28% of patients with multiple CNS AVMs have HHT. Some of these lesions will be micro AVM or micro AVF and superselective angiography is necessary to visualize them (figure 3) [6,7,8,9].

Certainly, there is a trend for presentation by the anatomic and physiologic character of the fistula. The term "hydrovenous disorder" aptly describes the relevant mechanisms operative in pediatric AVF. We owe a great deal to Dr. Lasjaunias who has described and coined this term. It has focused our attention to the venous side of vascular malformations and the dramatic effects elevated venous pressure can have on the developing brain.

Nearly all symptoms seen in AVF patients can be related to the venous side. With a high flow fistula, there is pressure transmission to the recipient veins. Several possibilities can occur. If there is a relative stenosis of the primary venous outflow, there can be rupture of the venous sac, enlargement and mass effect (as evidenced by skull erosion or cranial nerve effects, or very local brain venous hypertension resulting in seizures or focal deficit. If the fistula has no outlet stenosis until the venous sinuses exit the brain, then systemic venous pressure elevation results in impaired CSF resorbtion. Depending upon the age of the patient, there can be ventricular or extraventricular CSF space enlargement, increasing head circumference (before skull fusion), and eventually there will be venous congestive brain injury typified anatomically by calcification of the white matter at the grey-white junction (figure 2). The clinical correlates are headache, developmental delay, and/or seizures [10,11].

Haemorrhage in AVF patients can be explained by multiple mechanisms. With a proximal outlet stenosis, there may be inability of the venous wall to compensate with eventual rupture [12]. With a stenosis placed a little more distal in the outlet pathway, there can be focal or regional venous hypertension and bleeding

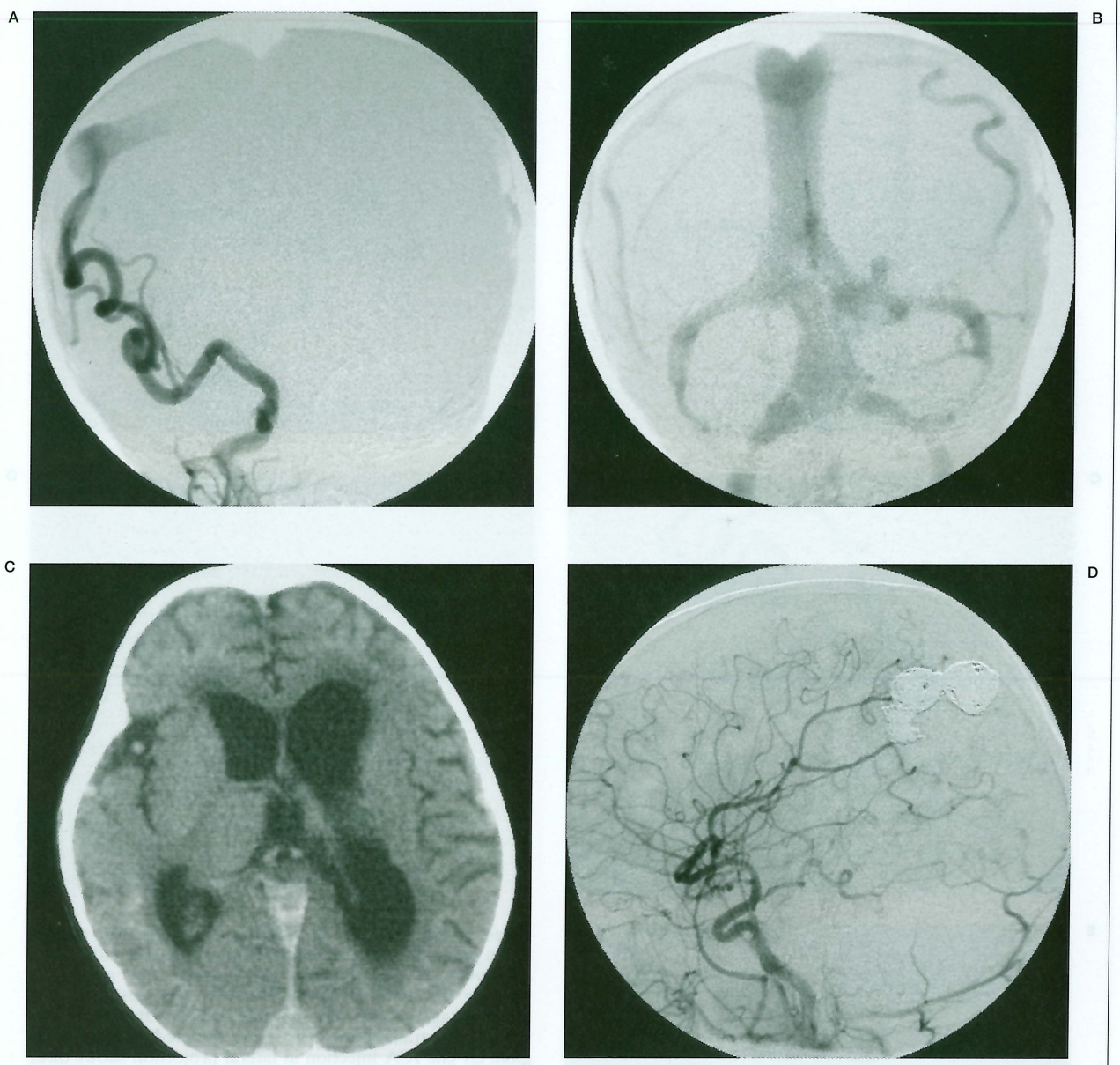

Figure 2 This is a 20 month old male who presented with developmental delay and seizures. This case demonstrates the consequences of the hydrovenous disorder. A) Right ICA angiogram pre-embolization shows the large fistula. B) ICA injection in ap view at the venous phase shows bilateral transverse sinus occlusion but with some outflow via the occipital/marginal sinuses but with cortical veins filling retrograde. C) CT scan showing ventriculomegaly with low density of the adjacent white matter at frontal horns and white matter calcifications in the temporal-occipital regions. D. ICA lateral post embolization using coils and glue (the fistula was initially fed by the middle, anterior and posterior cerebral arteries) without residual feeders. He has since advanced on milestones but still has some mild developmental delay.

of the adjacent brain whose venous outlet is impaired. With sinus outlet stenosis, the entire brain is involved with venous hypertension and remote areas may be subject to haemorrhage. [13,14,15,16] In one series, a further explanation was the presence of an inflow aneurysm as the putative cause of the haemorrhage [16].

Therefore, it is expected that larger, higher flow lesions become symptomatic in childhood. If the venous outlet has absolutely no stenosis, then the shunting leads to congestive heart failure in the neonate. If there is some stenosis, this initial diagnosis may not be made until there is haemorrhage, macrocrania or the subsequent

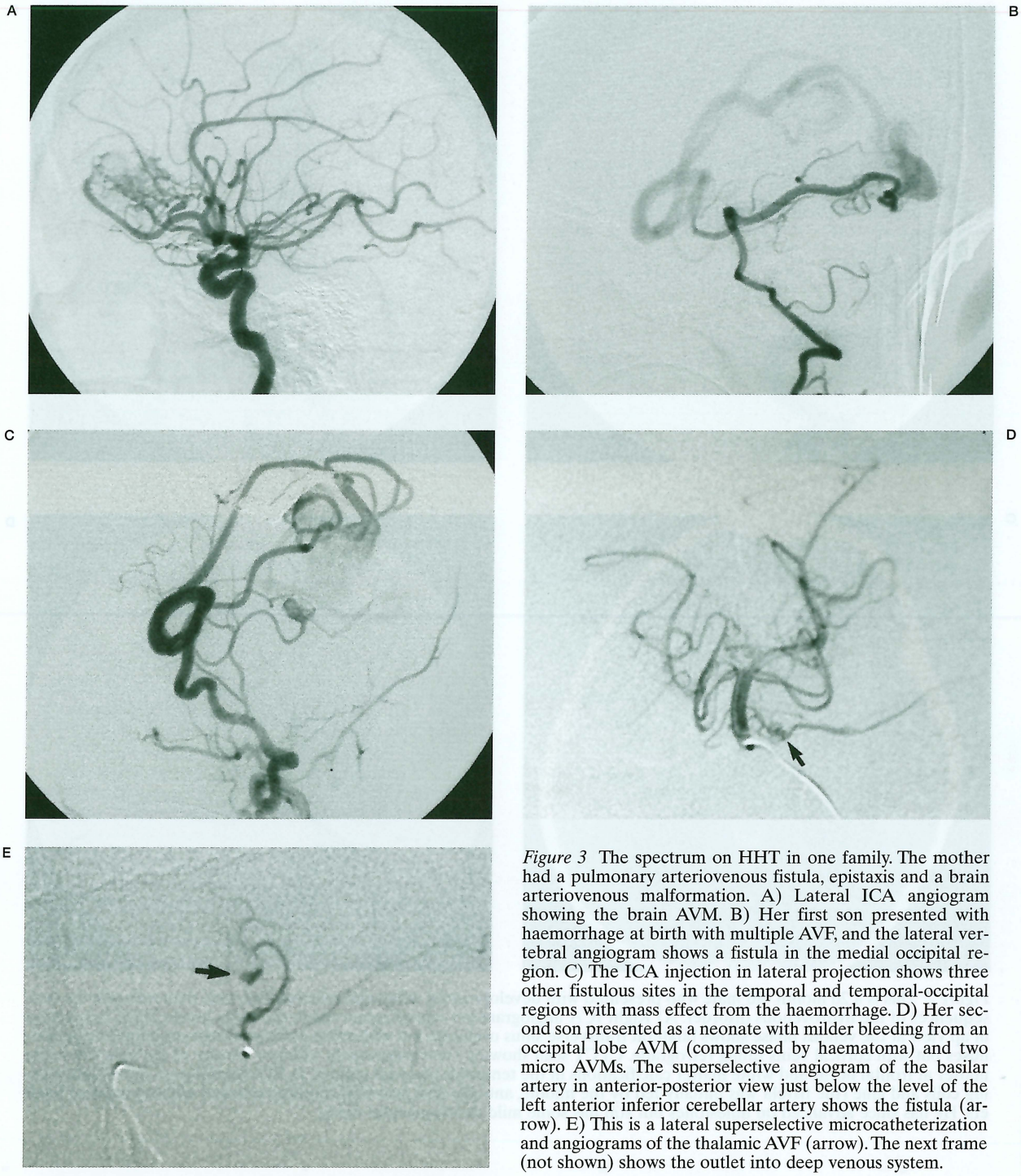

Figure 3 The spectrum on HHT in one family. The mother had a pulmonary arteriovenous fistula, epistaxis and a brain arteriovenous malformation. A) Lateral ICA angiogram showing the brain AVM. B) Her first son presented with haemorrhage at birth with multiple AVF, and the lateral vertebral angiogram shows a fistula in the medial occipital region. C) The ICA injection in lateral projection shows three other fistulous sites in the temporal and temporal-occipital regions with mass effect from the haemorrhage. D) Her second son presented as a neonate with milder bleeding from an occipital lobe AVM (compressed by haematoma) and two micro AVMs. The superselective angiogram of the basilar artery in anterior-posterior view just below the level of the left anterior inferior cerebellar artery shows the fistula (arrow). E) This is a lateral superselective microcatheterization and angiograms of the thalamic AVF (arrow). The next frame (not shown) shows the outlet into deep venous system.

injury to the developing brain (seizures, developmental delay). If the patient survives into adulthood, then there must have been sufficient compensation of the hydrovenous disorder or sufficient stenosis to prevent it. These patients then present with haemorrhage, focal signs (due to mass effect or regional venous hypertension), and headache.

This information points to the rationale for performing treatment. Although some fistulas are benign in their course, most require treatment.[4] In adults, haemorrhage is common and

careful analysis of the venous side is vital for treatment selection [5,12,17]. In pediatric cases, the treatment is even more urgent. Although there is some risk of haemorrhage, the primary issue is either congestive heart failure (in neonates) or the brain injury that occurs with the elevated venous pressures [18,19].

Treatment is aimed not only at the anatomic abnormality but also at the specific physiologic presentation. For those patients with CHF, the urgent process is cardiac stabilization, so flow volume reduction is the goal. For those with venous outflow disorder, if may be necessary to reduce arterial input and increase venous outflow.

The latter is a difficult proposition and little data exist on the durability of venous angioplasty and stenting in these patients, but some initial data on pseudotumor and dural fistula patients may be a guide to success with this therapy [20,21,22,23]. If haemorrhage or focal deficit is at issue, then mass effect reduction and complete eradication of the haemorrhagic source is necessary.

The complexity of the AVF is relevant to outcome, as accurate placement of the occlusion is necessary to assure cure before the venous outflow is compromised. There can be multiple connections at the arterio-venous transition that will not be addressed if blockage is done proximally in one of the arterial feeders. However, sequential blockage can be done do reduce flow prior to complete eradication (see figure 2) [5].

The major complication of treatment is haemorrhage. This can occur by two means. The first is occlusion of the venous outflow without eliminating the arterial input. The second is due to regional effects of the spontaneous venous thrombosis induced by the fistula occlusion. The conversion of the high flow patulous veins into slow flow/stagnant veins can lead to thrombosis. If there is venous drainage connection with the normal brain outlet, then that part of the brain will develop elevated tissue pressure, leading to haemorrhage. This is typically not an immediate phenomenon, but delayed after the embolisation [13,14].

In all cases complete AVF occlusion is the ultimate goal, but due to the complexities of the clinical or anatomic condition, this may not be achievable in one session. However, aggressive therapy for early onset presentation is warranted.

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